Chapter 14: Cardiovascular System

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A. Shock

Definitions

Shock

– Insufficient organ perfusion to meet tissue metabolic needs, leading to tissue hypoxia, acidosis, metabolic derangements and cell death

Hypotension:

- BP lower than expected range for age
- Hypotension does not necessarily mean that an infant is in shock

– There is no BP threshold below which intervention to increase BP has been shown to improve outcomes

– A combination of low BP with clinical signs of poor perfusion appears to be more strongly correlated with poor outcomes

Symptoms

• Tachycardia, poor perfusion/weak pulse, cold extremities, lethargy, apnea, tachypnea, metabolic acidosis

Classification of Shock

• Hypovolemic Shock
  – From blood loss-antenatal or postnatal
  – Post-operative due to capillary leak and third spacing of intravascular volume
    o Can also be seen in sepsis
• Drug Induced Hypotension
  – Magnesium sulfate, beta blockers (Labetalol), nitroprusside, narcotics, barbiturates
• Cardiogenic Shock
  • Cardiac failure – impaired filling, ventricular emptying, and/or contractility

–Birth asphyxia, CHD, metabolic abnormalities, arrhythmia, cardiomyopathy, obstruction to venous return

- Distributive shock (including septic shock)
  - Inadequate relative intravascular volume secondary to vasodilation
  - Septic shock due to release of endotoxins which lead to vasodilation
  - Also have capillary leak with third spacing due to endothelial injury
    - Anaphylaxis
    - Vasodilators
    - Adrenal insufficiency
- Neurogenic Shock
  - Birth asphyxia and IVH
- Shock in extreme prematurity
  - Due to hypovolemia, inability to regulate vascular tone, immature catecholamine response, IVH, adrenocortical insufficiency
  - Usually respond better to inotropes than to volume administration
  - PDA can cause transient hypotension

tages of Shock

Stage	Pathophysiology	Mechanisms	Change in vitals/lab values
Compensated	Heart, brain, lungs, kidney perfusion maintained, reduced flow to less vital organs	Vasoconstriction stimulated by acidosis/catecholamine release/decreased stimulation of baroreceptors à decreased urine output	Tachycardia -Stable BP -Normal HCO3 and lactate
Uncompensated Reversible	Decreased perfusion to all organs	Continuation of the above	- Increased tachycardia-BP begins to fall -HCO3 - decreases Lactate increases

Uncompensated Irreversible	Cellular dysfunction and acidosis secondary to ischemia à cellular death	Release of cellular mediators that lead to further reduced perfusion, injury to the endothelium, activation of coagulation cascade	Extreme tachycardia à bradycardia -Severe decrease in BP -Severe decrease in HCO3 -Severe increase in lactate

Diagnosis

- CBC with differential
- Blood culture
- ABG, lactate
- Electrolytes, glucose, calcium
- Newborn transfusion work-up
- Chest x-ray, echocardiogram, and head ultrasound

Treatment

- Treat underlying abnormalities
- To improve hypotension:
  - Volume expansion
- Normal saline bolus 10 ml/kg over 10-30 minutes
- Consider blood products for volume expansion
  - Low hematocrit
  - Bleeding
  - Electrolyte abnormalities that may be sensitive to additional dextrose or sodium
- Colloids associated with increased mortality
- May worsen cardiogenic shock
- Medications for hypotension

Medication	Dose	Mechanism	Adverse Effects	Notes
Dopamine	1-5 mcg/kg/min	Dopamine receptor Increases renal blood flow	Tachycardia, arrhythmias, tissue ischemia (only use in central IV)	Preferred inotrope in neonates esp. for <1500 gm. ++Chronotrope +Inotrope SVR effect is dose dependent
	5-15 mcg/kg/min	Dopamine and β1 and α receptors		
	15-20 mcg/kg/min	α receptors Systemic vasoconstriction		
Dobutamine	2-20 mcg/kg/min	β1>>β2 Increase contractility, decreases SVR +Chronotrope +Inotrope	Tachycardia, hypotension with hypovolemia, cutaneous vasodilation, arrhythmia, tissue ischemia	Better than dopamine in presence of myocardial dysfunction Less effect on heart rate
Epinephrine	0.1-0.3 mcg/kg/min	β1 & β2 Vasodilation, Increases contractility	Hyperglycemia, tachycardia, increased lactate, arrhythmias, tissue ischemia, hypokalemia	Most potent vasopressor ++Chronotrope +Inotrope
	0.3-1 mcg/kg/min	α receptors Vasoconstriction, increases HR		

Hydro-cortisone	Stress dosing: 1 mg/kg/ dose q8 hr. Physiologic dosing = 1 mg/kg/day q8-12 hr.	Increases the expression of adrenergic receptors in the vascular wall enhancing vascular reactivity to other vasoactive substances	Hyperglycemia, GI perforation/ hemorrhage, infection, cardiac hypertrophy	Use for unresponsive hypotension Do **not** use with indomethacin
Vasopressin	0.01 hr – 0.04 units/kg/hr	Vascular effects via G protein coupled V1a (vasoconstriction via IP3 pathway) and V2 receptors (vasodilation via cAMP) in cardiovascular system	Hypertension, electrolyte abnormalities, fluid overload	Vasoconstrictive effects predominate in IV infusion Minimal chronotropic and inotropic effects
Milrinone	Loading dose 50 mcg/kg over 15 minutes Maintenance 0.3-0.75 mcg/kg/min	PDE-3 inhibitor → increased intracellular cAMP, increased myocardial intracellular calcium, and increased uptake of calcium after systole	Hypotension, arrhythmias	Dosing extrapolated from older infants and children May potentiate diuretic effects Does **not** increase myocardial oxygen consumption

B. Hypertension (see also Chapter 18, Neonatal Kidney)

Definition

- Systolic/diastolic BP >95th percentile in right upper extremity
  - Term infant >90/60
  - Preterm infant >80/50

Etiologies

- Renal artery or aortic thrombosis
- Primary renal disease
- Obstructive uropathy
- Coarctation of the aorta
- Endocrine disorders: hyperthyroidism, CAH (11-betaOH)
- Medications: theophylline, corticosteroids, pancuronium
- BPD
- Pain, agitation, drug withdrawal

Diagnosis

- Four extremity BPs-evaluate for coarctation
- Labs
  - UA, Urine culture
  - Urine Protein / Urine creatinine (Normal <1)
  - Electrolytes, creatinine, BUN
  - Plasma renin activity, aldosterone
  - TSH, free T4
- Imaging
  - Abdominal/Renal ultrasound with Doppler studies
  - Echocardiogram

Treatment

- Nephrology consult to determine appropriate medication
  - Usually start with a calcium channel blocker (isradipine)

C. Arrhythmias Complete

Heart Block

- Seen with maternal connective tissue disorders (i.e. SLE) who have anti-SSA (Ro) or anti-SSB (La) antibodies
  - Can lead to hydrops fetalis
- Treatment
  - Only necessary if symptomatic
  - Generally symptomatic if HR < 55 bpm
  - Atropine, isoproterenol, pacemaker

Supraventricular Tachycardia

- HR 230-330 bpm with decreased variability (fixed R-R interval)
- Increased risk with CHD (Ebstein's anomaly, L-TGA), WPW
- Acute Treatment
  - Unstable-synchronized cardioversion
    - Start with 0.5 J/kg, increasing by 0.5 J/kg to max 2 J/kg
  - Stable
    - Vagal maneuvers-gag reflex, ice to the face, knees to chest
    - Adenosine
      - 50 mcg/kg rapid IV push followed by rapid saline flush
        - Via PIV with 3 way stop cock for rapid flush
      - Increase by 50 mcg/kg every 2 minutes to max dose of 250 mcg/kg
      - Causes transient AV node block-have ECG running and defibrillator nearby

D. Congenital Heart Disease (CHD)

- VSD-most common CHD
- Transposition of the great arteries-most common CHD presenting in the first week of life
- HLHS-second most common in the first week of life and the most common cause of mortality in the first year of life
- Tetralogy of Fallot-most common CHD presenting after the first week of life

Diagnosis

- Four extremity blood pressures

- Pre-and post-ductal O_2 saturations (CHD screen)
- Chest x-ray-evaluate heart size and pulmonary vascular markings
- ECG
- ABG-evaluate for metabolic acidosis and hypoxemia
- Echocardiogram

Clinical Presentation

- Respiratory Distress: VSD, PDA, ASD, TAPVR, truncus arteriosus (TA)
- Murmurs
  - Systolic
    - Holosystolic – VSD
    - Ejection – aortic/pulmonic stenosis or obstructed outflow tract
    - Click – aortic/pulmonic stenosis or truncus arteriosus
  - Blowing
    - Valve regurgitation
  - Diastolic *always pathologic
    - Aortic/pulmonic regurgitation, tricuspid/mitral stenosis, increased flow across tricuspid/mitral valves
  - Continuous
    - PDA, AV fistula, venous hum, collateral vessels, truncus arteriosus, aortopulmonary window
  - Gallop
    - Decreased ventricular compliance and high-flow states
- Cyanosis – bluish discoloration of the tissues when deoxygenated hemoglobin in the capillary >3g/dL
  - Appearance of cyanosis depends upon the total amount of deoxygenated hemoglobin, not ratio of deoxygenated to oxygenated blood
  - Cyanosis with normal or increased pulmonary blood flow: TGA, TA, DORV
  - Cyanosis with decreased pulmonary blood flow: TOF, tricuspid atresia, pulmonary atresia/stenosis, Ebstein's anomaly
  - Differential Cyanosis - >10% difference in pre/post-ductal saturations
    - Lower body more cyanotic than upper body – R to L ductal shunting with increased PVR
    - Seen in coarctation of the aorta, pulmonary hypertension, interrupted aortic arch

- Reverse differential cyanosis
  - Upper body more cyanotic than lower body
  - Seen with dTGA + coarctation of the aorta, pulmonary hypertension, or interrupted aortic arch
- Shock: TAPVR with obstruction, HLHS, critical aortic stenosis, interrupted aortic arch, coarctation of the aorta

Management

- IV access; UAC, UVC, PICC line
- Prostaglandin E1
  - –For ductal-dependent lesions
  - –Dose: start at 0.01-0.02 mcg/kg/min for known ductal dependent lesions or lesions presenting soon after birth
    - If presenting several days after birth, consider starting at 0.5-1 mcg/kg/min
  - –Side effects: apnea (may be treated with caffeine), fever, leukocytosis, cutaneous flushing, bradycardia, hypotension, hypoglycemia, hypocalcemia
  - –Long-term causes reversible cortical proliferation of the long bones, and gastric outlet obstruction
- Generally avoid supplemental oxygen as this causes pulmonary vasodilation and will increase pulmonary blood flow at the expense of systemic blood flow
  - –Maintain oxygen saturations around 75-80% = Qp/Qs of 1
  - –$Qp/Qs = (SaO_2\text{-}SvO_2)/SpvO_2\text{-}SpaO_2)$
    - Ratio of pulmonary to systemic blood flow
- Cranial ultrasound, renal ultrasound
- Genetic testing

E. Patent Ductus Arteriosus in Preterm infants

Clinical Presentation

- Murmur-LUSB, systolic or continuous
- Hyperactive precordium, bounding pulses, palmar pulses
- Widened pulse pressure (>30 mmHg)
- Worsening respiratory distress

- Hepatomegaly, cardiomegaly
  - –Neither individual clinical trials nor meta-analyses have demonstrated that closing PDA results in improved long-term outcomes in preterm infants
  - –Trend toward a more conservative approach to PDA management

PDA Guideline UnityPoint-Meriter and AFCH

PDA Treatment Guideline

Timing of Initial echocardiogram

- 22 0/7- 25 6/7 weeks or <750g: Obtain a routine echocardiogram on day 3-5
- 26 0/7- 28 6/7 weeks: Obtain an echocardiogram on day 7 or after if clinical score is >=3
- Discuss with the Cardiology team about potential limited echo if the infant is critically ill

Medical Treatment of PDA

- Medical treatment is indicated if McNamara Echocardiographic score is >=3
- Decision for subsequent treatment courses based upon clinical judgement if echocardiographic score >=3
- Total of three courses of medical treatment is recommended
- Choice of medication based on provider preference and clinical status of the patient
- Medication and dosing
  - Ibuprofen: 10 mg/kg NG x 1, then 5 mg/kg q24h x 2 more doses NG
    - Do not use if evidence of renal dysfunction; SCr >1, AKI in past 7 days (rise of SCr by 0.3 or UOP < 0.5 mL/kg/d)
    - Do not use if GI bleeding or platelets < 100K
    - Do not use if hydrocortisone administration within 24 hours
  - Acetaminophen: 15 mg/kg NG q6h x 5 days
    - Do not use if evidence of liver injury/ cholestasis
  - Use IV ibuprofen or IV acetaminophen if on < 60 ml/kg/day of feeds
- Lab monitoring prior to each course:
  - Ibuprofen: BMP and platelets
  - Acetaminophen: nutrition panel
- No need to reduce or withhold advancing feeds while on medical treatment for PDA

Definitive Closure of PDA (Transcatheter closure/ PDA Ligation)

- If combined score is >=7 after three courses of medical treatment or if medical treatment is contraindicated, consult Pediatric Interventional Cardiology to discuss definitive PDA closure
- Preferred time for transcatheter closure of hemodynamically significant PDA is 21-35 days

Table. Modified McNamara Scale

Points	Clinical Score	Echocardiographic Score
1	RSS < 1.5	Continuous flow and increasing velocity flow into the branch PAs: <0.15 m/sec in diastole in LPA
2	RSS 1.5 – 1.8	Small PDA, Continuous flow and increasing velocity flow into the branch PAs : >0.4 m/sec in diastole in LPA
3	RSS 1.8 – 3.0 OR hypotension requiring a single vasopressor	Moderate PDA, Diastolic flow reversal in the descending aorta below the level of the PDA
4	RSS >3.0 OR hypotension requiring more than 1 vasopressor	Large PDA, A dilated LA (typically 2 times larger the aorta in PLAX view)
5		Large PDA, LV dilation

RSS – Respiratory Severity Score (MAP x FiO2)

F. Persistent Pulmonary Hypertension Diagnosis

- Pulmonary hypertension should be considered in a term/post-term infant with cyanosis
- Associated with fetal distress, RDS and meconium aspiration syndrome
- Pre-ductal and post-ductal saturations differ significantly (>10%)
- Desaturation with stimulation, crying
- S2 is loud with diminished split, murmur of tricuspid regurgitation
- Chest x-ray-decreased pulmonary vascular markings
- Echocardiogram
  - –PDA with R $\rightarrow$ L shunting
  - –Flattening of the interventricular septum
  - –Bulging of the atrial septum
  - –Pulmonary pressures determined using TR velocity

Treatment

- Minimize handling
- Surfactant-for RDS or meconium aspiration
- Sedation/paralysis
- Supplemental oxygen as needed to maintain saturations within goal range: Dilates pulmonary vasculature
- Correct acidosis: Acidosis leads to pulmonary vasoconstriction
- Inhaled Nitric oxide: See Respiratory Chapter
- Sildenafil
- Inotropic agents to increase systemic pressures (decreasing shunting)
- ECMO – consider if oxygenation index (OI) is > 35 for 5-6 hrs

$$OI = \frac{\text{mean airway pressure} \times FiO_2 \times 100}{PaO_2}$$

G. Miscellaneous

Electrocardiogram

- Differs from the adult
- RV dominance with right axis deviation
- T wave inversion in V1-V4 after 48-72 hours of age is normal
  - –If T wave inversion is not present consider RVH
- RSR' in right precordial leads is normal as long as QRS interval is <10 msec over normal intervals
- QTc interval
  - –QT/square root of the previous R-R interval
  - –<0.47 normal in first week of life
  - –<0.45 normal from 1 week to 6 months of age

Equations

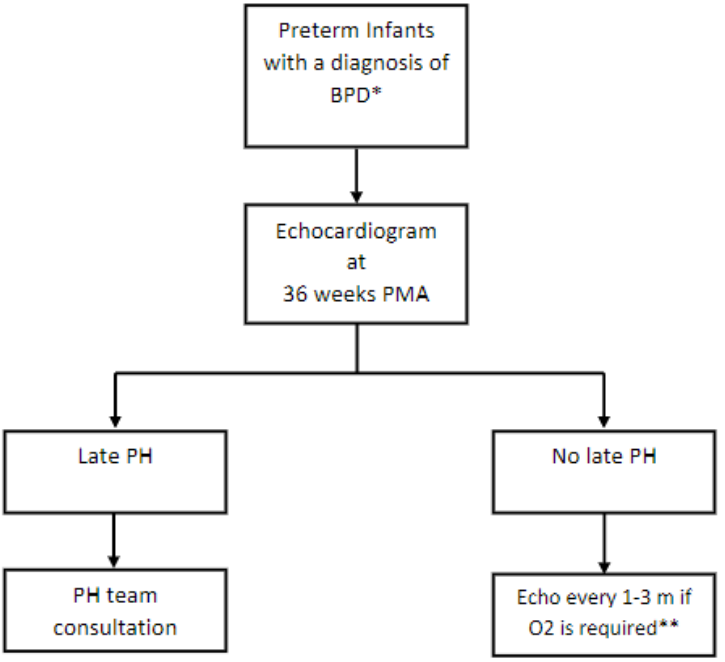
- Shortening Fraction $= \frac{\text{LV diastolic diameter} - \text{LV systolic diameter} \times 100}{\text{LV diastolic diameter}}$
  - –Normal is 28-40%
- Ejection Fraction $= \frac{\text{LV end-diastolic volume} - \text{LV end-systolic volume} \times 100}{\text{LV end-diastolic volume}}$

- Cardiac Output = Stroke volume x Heart rate

–Stroke volume is affected by preload, afterload, and contractility

–In the neonate the CO is more dependent on heart rate

Pulmonary Hypertension Screening guideline for preterm infants



*BPD defined as need for respiratory support at 36 weeks PMA

**Every 1-2 months if inpatient, every 3 months if following up outpatient

Indicate "Evaluate for pulmonary hypertension" on the echo request.

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